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CASE REPORT

Neonatal Naso-Oro-Hypopharyngeal Langerhans Cell Histocytosis: Case Report

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Abstract

Langerhans cell histiocytosis (LCH) is a rare histiocytic disorder most commonly characterized by single or multiple osteolytic bone lesions demonstrating infiltration with histiocytes having bean-shaped nuclei on biopsy. These histiocytes, along with lymphocytes, macrophages, and eosinophils may infiltrate nearly every organ (most notably the skin, lymph nodes, lungs, thymus, liver, spleen, bone marrow, or central nervous system with the exception of the heart and kidneys). [(1)] We, hereby, describe a rare case of Naos-Oro-Hypopharyngeal *LCH* in a neonate.

Keywords: Langerhans Cell Histocytosis, Naso-Oro-Pharyngeal, Neonatal, Oncology, NICU, Paediatric, Saudi Arabia.

List of Abbreviations	
LCH	Langerhans cell histiocytosis
NICU	Neonatal Intensive Care Unit
HFNC	High Flow Nasal Cannula
PDA	Patent Ductus Arteriosus
ASD	Atrial Septal Defect
LVH	Left Ventricular Hypertrophy
E.N.T	Ear Nose and Throat

1 | INTRODUCTION

Langerhans cell histiocytosis (LCH) encompasses a heterogenous group of diseases, including histiocytosis X, eosinophilic granuloma, Letterer–Siwe disease and many more which share a common feature of Langerhans cell proliferation. Clinically, patients affected present differently in highly variable fashion. Noting that, severity and prognosis of the disorder are closely correlated to the type and extent of organ involvement. [2]

Several theories have been proposed to explain the pathophysiological basis of LCH. These include uncontrolled immunologic stimulation of a normal Langerhans cell resulting in the proliferation and accumulation of these cells, histiocytic reaction secondary to infection or excessive cytokines or lymphokines production and neoplastic proliferation of monocyte-macrophage cell's series. [3]

So far, we have caught sight of a unique presentation in an unusual site and age group as we describe the biopsy-evident skin-free mass-presented neonatal LCH of naso-oro-pharyngeal territory.

2 | CASE REPORT FEBRUARY / 2021

A presently 2-month-old baby girl, who was born at term (40 weeks), and occurred to be a product of cesarean section due to an antenatal finding of polyhydramnios ultrasonically.

At birth, she weighed 3600 grams and her APGAR score was nursery-acceptable initially, yet shortly after delivery, she experienced respiratory distress owing to copious amount of nasal and oral secretions in conjunction with stridor. Soon after, she was intubated for 24 hours and nictitated to be entertained in the NICU. She was weaned gradually down to the mode of HFNC at 6 litres of oxygen.

In the light of her persistent respiratory discomfort and the questionable possibility of ? Tracheoesophageal fistula, she was subjected to a naso-pharyngeal Computed Tomography with echocardiography, which demonstrated left-sided choanal membranous stenosis and small PDA with left-to-right shunt ASD and mild LVH respectively.

E.N.T services were then sought as she worsened for respiratory compromise which rendered her intubated again, they in return, advised for a CT repeat of the naso-pharynx and neck to rule out congenital malformation. It revealed ‘’naso-pharyngeal mass measuring 3.5 x 2 x 2.5 cm with enhanced calcifications and cortical bone erosions of the clivus imposing mass effects on the airway", a confirmatory Magnetic Resonance Imaging was performed and concluded ‘’Large nasopharyngeal/clival mass causing obstruction of the nasopharyngeal airway with evidence of skull base involvement" awaiting biopsy diagnosis for 60 days, when she was transferred for a higher centre for the procedure.

Interestingly, the excisional biopsy showed ‘’an infiltrative lesion composed of epithelioid cells with abundant pale eosinophilic cytoplasm with irregular nuclei and prominent nuclear grooves associated with few eosinophils in the background. This morphology along with the immunohistochemical profile is consistent with Langerhans cell histiocytosis".

In the light of this, Chemotherapy was commenced and subsequent good response was observed.

Respective relevant Labs and imaging are stated and illustrated below in (Table 1), (Figure 1) respectively:

Supplementary information The online version of this article (https://doi.org/10.15520/jmrhs.v4i3.330) contains supplementary material, which is available to authorized users.

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TABLE 2: Laboratorywork-up.

Labs/Imaging / Date	Present day	Reference Range
WBC	6.3 *10e9/L*	4.3 -11.3 *10e9/L*
ANC	3.38 *10e9/L*	1.35 – 7.5 *10e9/L*
Hemoglobin	13.3 *g/dl*	11-15 *g/dl*
Platelets	829 *10e9/L*	155 - 435*10e9/L*
Reticulocytes	0.14 %	0.5 - 1.5 %
ALT	24 *U/L*	0 – 55 *U/L*

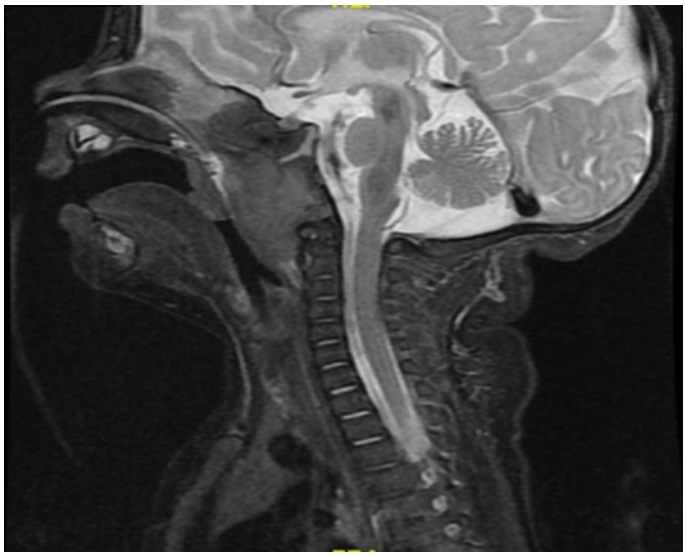

FIGURE 1: MRI of the nasopharynx and neck.

3 | DISCUSSION

LCH diseases are usually affecting children and involving one or many body systems including head and neck although invoving paranasal sinuses is rare. Biopsy and histopathological examination is core in diagnosis of LCH which shows multinucleated Langerhans' cells, histiocytes, and eosinophils.

Our reported case have spotted the light on recognizing the entity of cutaneous-spared LCH in neonatal period. Although head and neck are considered the most commonly involved area in LCH, mass extension throughout the whole upper airway is distinctly rare as described above.

There were multiple reports on LCH presentation in relatively uncommon areas of the body, namely ''Mandible, Maxillary sinuses, tonsils, etc.." (2) , (3) , (4)

Treatment is controversial in multi-organ disease. Several studies have shown that multi-agent chemotherapy have overweighted result compared to high dose prednisolone alone in matter of better response rate and less recurrences specially if given in longer duration. (3)

Children younger than 2 years old and the presences of organ dysfunction and multi organ involvement specially involvement of high-risk organs (liver, spleen, lung and bone marrow) found to be associated with poorer prognosis in addition of response to initial therapy (assessed after 6-12 weeks of treatment). (3)

The overall survival rate for all was 79% at 1 year, 74% at 3 years, and 71% at 5 years and the outcome for children with LCH involving low-risk organs has always been excellent with 20% - 30% incidence of relapse. (3)

Nevertheless, further observations are needed to examine the prevalence of these unusually affected body sites.

4 | CONCLUSIONS

LCH is a rare multisystemic with a characteristic infiltration of most of the body organs, notably the skin, lymph nodes, lungs, thymus, liver, spleen, bone marrow, or central nervous system. However, we came to remark a rare form of LCH involving the upper airway exclusively. We, hereby, urge physicians to be enlightened in regards to this presentation.

ETHICAL CONSIDERARTIONS & IRB APPROAVAL

A written consent was obtained and signed by the legal guardians of the baby for full disclosure while maintaining strict confidentiality in respect to the patient medical information and images under the approval of King Fahad Medical City (KFMC) research centre ethical committee. Under the umbrella

of KFMC Institutional Review Board (IRB), this case was approved.

DATA AVAILABILITY

The data that support the findings of the study are available from the corresponding author upon reasonable request.

CONFLICT OF INTEREST All authors have no example conflicts of interest to disclose.

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AUTHORS' CONTRIBUTION ZU; The primary author, attending physician and reviewing the whole manuscript.

AE ; Co-author, Planning the case report conception and design, reviewing relevant literature and the final writing.

MS & AH; Co-author, Writing the case report scenario.

AW; Co-authors, Reviewing and writing the Labs.

MQ; Co-author, Reviewing discussion.

MG; Co-author, Reviewing conclusion, references and pictures.

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